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Solitary Fibrous Tumour of the Retroperitoneum Mimicking a Renal Mass

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Authors report a case of solitary fibrous tumour of the retroperitoneum that mimicked a renal mass. A review of the literature and a discussion on the biologic meaning of the lesion are presented.

Solitary fibrous tumour is the name generally given to a neoplasm that most frequently occurs in the pleura [1–3].

The occurrence of such a neoplasm at sites other than the pleura had been sporadically reported [4–6], with a very few occurring in the abdominal cavity [7, 8].

We report here a case of retroperitoneal solitary fibrous tumour that caused considerable diagnostic problems due to its unusual site of origin.

Case report

A 51-year-old Caucasian woman was referred to our Institute for evaluation of a left renal mass in a single functioning kidney discovered during a screening for renal failure. Her past medical history revealed that she was subjected to right nephrectomy for tuberculosis in 1972. In 1990 she was again evaluated by us for symptoms suggesting early painful bladder disease; she underwent bladder distension with definitive improvement. By that time the medical history was mute until the present problem.

On admission creatinine was 134 μmol/l (n.v.: 44–115 μmol/l), Na 141 mmol/l (n.v.: 136–145 mmol/l), K 4.1 mmol/l (n.v.: 3.5–5.0 mmol/l), Cl 108 mmol/l (n.v.: 96–108 mmol/l). Urine examination was normal.

Ultrasonography performed elsewhere showed a hyperechoic renal mass about 6 cm in diameter.

She was then subjected to a CT scan that demonstrated a mass about 7 cm in diameter extending from the renal pelvis and involving the renal parenchyma with post-contrast enhancement (Fig. 1). Surgical planning also included arteriography. The patient was then subjected to explorative laparotomy; this revealed an apparently capsulated, well circumscribed, firm mass involving the middle third of the kidney but easily dissectable from it (Fig. 2). The postoperative course was uneventful; on discharge creatinine was 144 nmol/l.

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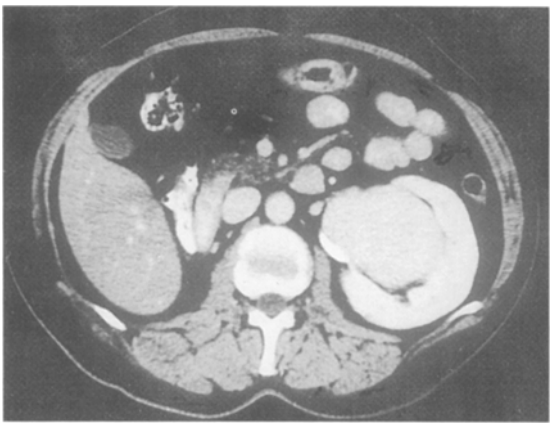

Fig. 1. CT shows a mass of 7 cm extending from the renal pelvis and involving the parenchyma with post-contrast enhancement

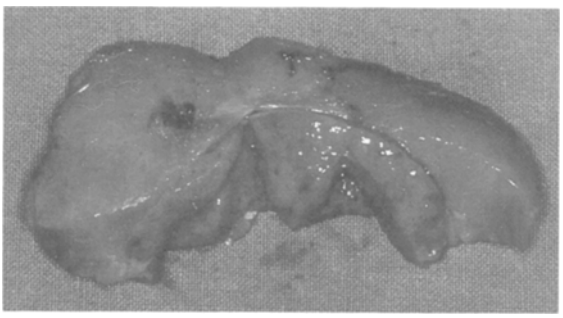

Fig. 2. Surgical specimen

Histology

The lesion presented a cellular density area greater at the periphery, with pleomorphic round/ovalar and spindle cells, scanty cytoplasm in a myxoid component of collagenized and haemangiopericytoma-like areas. We found no necrotic areas and mitoses were very rare (1×10 HPF) (Fig. 3). The immunohistochemical finding showed strong positivity to vimentin (HHF 35 was positive for the vascular areas); keratins, desmin, neuroendocrine marker (S-100) and EMA were negative.

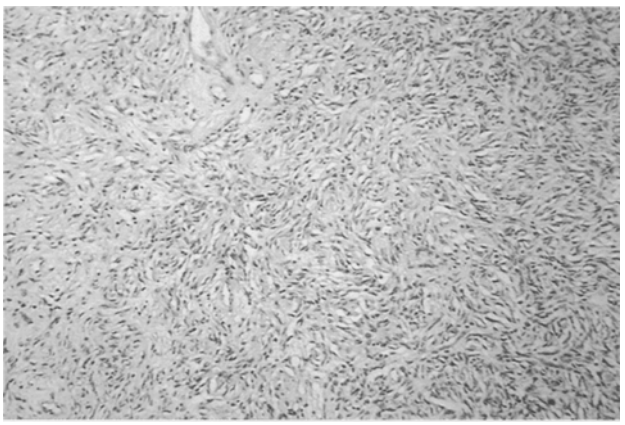

Fig. 3. Solitary fibrous tumour with haemangiopericytoma areas; the intervening stroma is infiltrated by fibroblasts with slight nuclear abnormalities. Absence of mitoses or necrosis (H&E, ×8)

Discussion

Solitary fibrous tumours are rather rare but otherwise well established neoplasms of the pleura [1–3]. However, the occurrence of such a tumour at other sites has been occasionally reported with the most common unusual sites being the lungs [1, 4], mediastinum [5], pericardium [9], nose [6], peritoneum [7, 8] and retroperitoneum [7, 8].

The rarity of the abdominal localization can easily explain the diagnostic challenge it presents to the clinicians. As far as we know, till now only 4 cases were located in the retroperitoneum [7, 8] and in no case has there been an involvement of the kidney.

Histologically the tumour may cause some problems in differential diagnosis from malignant mesotheliomas, fibrosarcoma, leiomyoma and leiomyosarcoma, but again the difficulty is due to the unusual location rather than the histologic features. Immunohistochemistry and histochemistry can help in the diagnosis; in the present case, according to the literature [7, 8, 10], immunohistochemistry showed strong positivity to vimentin; keratins, desmin, neuroendocrine marker (S-100) and EMA were negative. These findings are also of interest in considering the histogenesis of this neoplasm, currently reputed to originate from the submesothelial mesenchymal cells [7, 8 10]. The extrathoracic location is usually clinically mute even if symptoms related to size and location are reported; also of interest is that the tumour had been reputed to elicit hypoglycaemia as well as arthralgia and osteoarthropathy [2, 8].

The behaviour of this neoplasm is unpredictable: an aggressive behaviour with the possibility of local invasion, recurrence and even metastasis have been reported in large series of pleural tumours [2]; in the abdominal loca-

tion, the fibrous tumour never showed recurrence or spread even if the short-term follow-up available for most of these rare cases may play a role [7, 8]. In case of a large inoperable mass, radiotherapy has been shown to be of value [8].

Conclusion

Even if rare, the possibility of an extrapleural location of fibrous tumour must be borne in mind; it will make easier to set up the correct diagnosis for both the pathologist and the clinician. Symptoms and radiologic findings are not helpful in the diagnosis. In case of unresectable masses, external beam irradiation may be of value.

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